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CASE STUDY

Venous malformation of right angular vein

Malformación venosa de la vena angular derecha

Hemanth Vamanshankar*, **Jyotirmay Shyamsundar Hegde**, **Pradipta Kumar Parida**

Department of ENT, Jawaharlal Institute of Postgraduate Medical Education and Research, Pondicherry, India

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Case report

A 34-year-old lady presented to us with complaints of a progressively enlarging swelling in the region between the medial canthus of right eye and root of the nose, since 4 years. She did not give any history of nasal complaints, epiphora or pain from the swelling. There was no history of trauma to face in the past.

On examination, the swelling was found to be about 2 cm × 2 cm in size, soft in consistency and non-tender. There was a bluish hue of the overlying skin. The underlying bone was found to be intact. The swelling was completely compressible on palpation, and would slowly fill up on release. An increase in the size of the swelling was noted on performing valsalva manoeuvre.

X ray of the paranasal sinuses did not reveal any underlying bony defect. We proceeded to do a contrast enhanced computed tomogram (CT) in order to establish the diagnosis. CT revealed a 2 cm × 1 cm homogenously enhancing soft tissue density with normal surrounding bony and soft tissues. There was no surrounding soft tissue inflammation or collections noted. The above lesion appeared to be in continuous with a tortuous dilated angular vein which in turn drained into the right external jugular vein (Fig. 1). Hence a diagnosis of venous malformation was established.

This being a small lesion in the face, the patient was planned for a surgical excision of the lesion under local anaesthesia (Fig. 2A). A curvilinear incision was made over the lesion (Fig. 2B) and skin flaps were raised. On careful dissection, the ectatic venous lesion was found just above the underlying bone, and removed along with the underlying periosteum (Fig. 2C). The patient had an uneventful recovery period.

Histopathology of the specimen showed many dilated venous calibre intercommunicating vascular channel, with walls showing focal myxoid change and eccentric wall thickness, suggestive of a venous malformation (Fig. 3). The patient is free of any symptoms since 4 months after the surgery.

Discussion

Congenital disruption in the development of a normal vein results in the formation of venous malformations (VM). These soft tissue masses can accumulate static blood over a period of time, causing them to lose their vascular elasticity, and expand. Left untreated, they cause loss of function and disfigurement; may facilitate the development of a thrombus and can be painful.[1]

Venous malformations have an incidence of 1 in 10,000.[2] They are mostly sporadic in nature, but familial and multifocal patterns have been described. Chromosome 9p mutation has been implicated in some inherited forms of venous malformations.[3] Patients of VM's have been found to have upregulation of factors like tissue growth factor beta

* Corresponding author.
E-mail address: vhemanth2000@yahoo.com (H. Vamanshankar).

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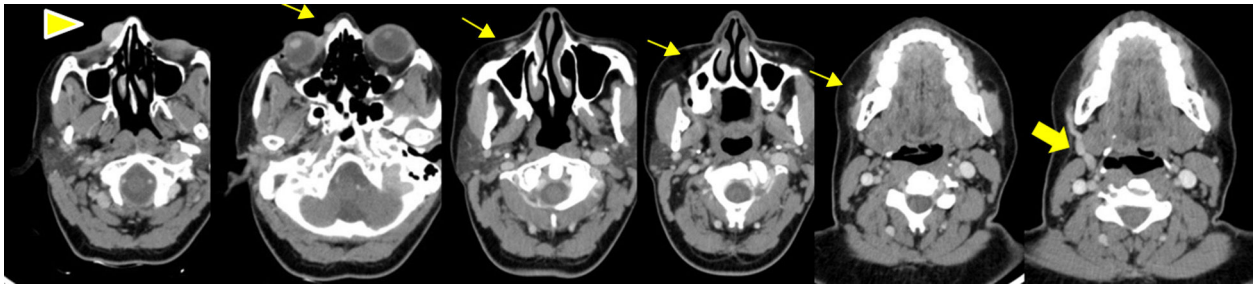

Figure 1 Serial contrast enhanced CT of the nose and paranasal sinuses showing the right tortuous dilated angular vein draining into the right external jugular vein (venous malformation shown with arrow head; angular vein course shown with small arrow; angular vein entering external jugular vein shown with large arrow).

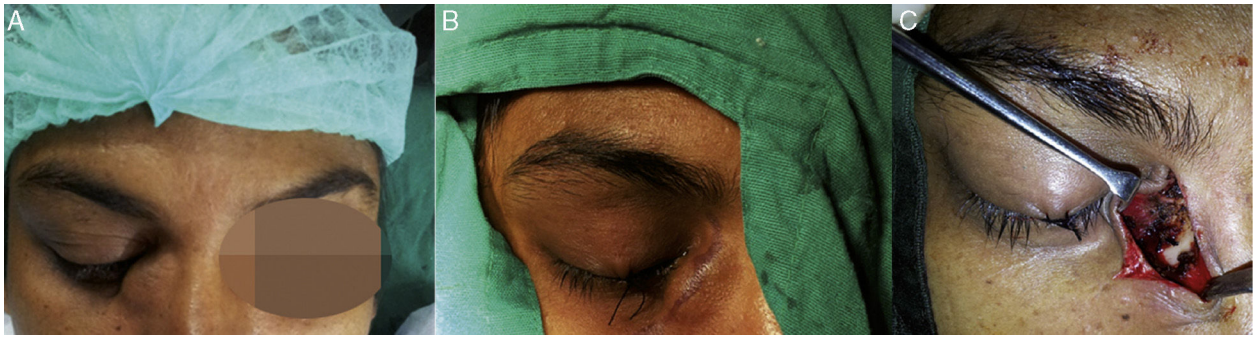

Figure 2 (A) Pre-operative photograph showing swelling between right medial canthus and root of nose. (B) Per-operative photograph showing the curvilinear incision made over the swelling. (C) Per-operative photograph – after excision of the mass – showing the bed of the lesion with intact underlying bone.

(TGF-beta) and basic fibroblast growth factor (beta-FGF)[4]; progesterone receptors have also been found accounting for the rapid expansion in these lesions during hormonal changes.[5] Mutation has also been found on the angiopoietin receptor gene TIE2/TEK in many sporadic forms.[6]

After hemangiomas and lymphatic malformations, they are the third most common vascular masses in the head and neck region. Frequently involved areas of the head and neck include the lips, tongue, aero-digestive tract and muscles of mastication.[1] The site of occurrence of our patient's swelling – medial to medial canthus of right eye is rare, and to the best of our knowledge, not been described in literature.

Venous malformations can present at birth, early childhood or in adults. They may present as a rapidly expanding, painful mass if a thrombus is formed. Others can present as a bluish painless subcutaneous mass, as in our patient. The

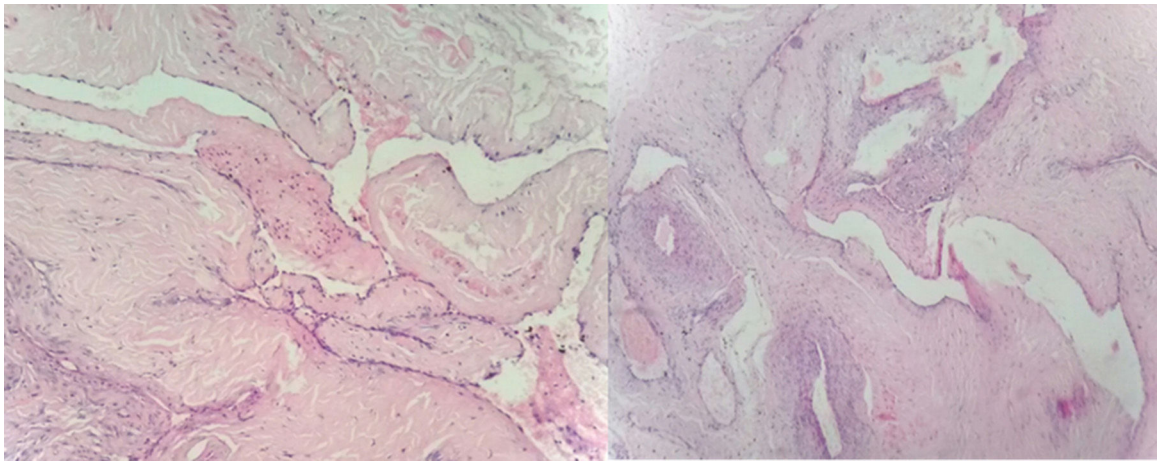

Figure 3 Haematoxylin and eosin stain of the specimen showing many vessels of dilated venous calibre with walls showing focal myxoid change and eccentric wall thickness (10×).

diagnostic hallmark of a venous malformation is expansion of the mass on performing valsalva manoeuvre – due to the dilatation of the ectatic veins, and the fact that they are compressible.[1] This was demonstrable in our patient.

Diagnostic modality of choice for venous hemangiomas is ultrasound.[7] MRI can be useful if the lesion involves multiple planes or in case of infiltrative lesions. They appear hyperintense on T2 and iso/hypointense on T1 weighted imaging.[8] D dimers may be elevated in large lesions and are markers of the disease.[9] Our patient was investigated with a CT scan.

Treatment modalities of venous ectasias can vary, according to the site and size of the lesion. Patients often seek treatment in view of aesthetic or functional impairment, chronic pain in swelling or due to its unrelenting growth. Treatments include laser photothermolysis, surgical excision, sclerotherapy or a combination of these.[1] Surgery remains one of the superior treatment options in localised forms, and may offer a cure.

Conclusion

Although rare, venous ectasias should be considered as a differential diagnosis in swellings of the head and neck region, even in the case of adults. A high index of suspicion and a thorough clinical examination helps in the diagnosis, which can then be confirmed radiologically. In the region of the head and neck, surgical modality of treatment still remains a superior option for venous malformations.

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Conflict of interest

None.

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